From: "ROOT" <root@sctimst.ac.in>
To: "ROOT" <root@sctimst.ac.in>
Date: 05/08/2024 01:10 PM
Subject: CPC Clinical Protocol 07.08.2024

Dear All,
Season's Greetings.
The next Wednesday CPC of the session will be held on **August 07, 2024** at 08.00 hours (IST) in **Lecture Theatre 1**, Nehru Hospital, PGIMER, Chandigarh.

The session will also be available on the Webex platform. Kindly follow the link below to join.
https://telemedicine.webex.com/telemedicine/j.php?MTID=mb8f5c93d0efe86e2ec7df721b3c21227

In case you join in thru WebEx, kindly **ensure that your microphone and camera are switched off** and **PLEASE DO NOT SHARE YOUR SCREEN**.

The Clinical handout of the case to be discussed is attached herewith.
The clinical protocol will be presented by ***Dr. Anju Gupta, Department of Pediatrics.*** Radiology will be presented by ***Dr. Vikas Bhatia***. Autopsy pathology will be presented by ***Dr. Debajyoti Chatterjee.***

Yours sincerely,

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Regional Resource Centre, North
Department of Telemedicine
PGIMER, Chandigarh

Staff CPC
07.08.2024

U, 4.5 months old boy	CR No: 202301308969	Clinician Incharge: Prof N Sankhyan
DOB 09.09.2022	Adm No: 2023007568	Clinical Discussant: Anju Gupta
DOA: 28.01.2023	R/O Amritsar	Pathologist: Dr Debajyoti Chatterjee
DOD: 18.02.2023		Radiologist: Dr Vikas Bhatia

HOPI

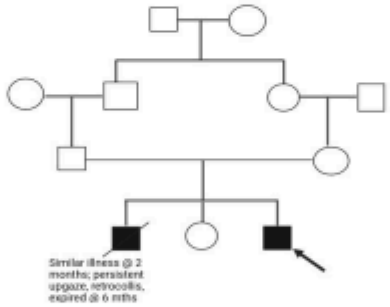
Baby was born at term/2.75 Kg/ by normal vaginal delivery. There was no birth asphyxia, hypoglycemia or jaundice after birth. He was breastfed on day 1 itself and had a good suck and swallow. He continued to have normal development till the age of 2.5 months with acquisition of milestones such as social smile, following objects and cooing. He could raise his head above ground when made to lie prone.

Parents started developing concern by 3.5 months of age when they noticed progressive insidious onset deterioration in the form of

- Thinness and ↓ movement of legs and arms
- ↓ interaction
- ↓ following of objects
- ↓ volume of cry
- Squint

Baby also started developing recurrent cough with pooling of secretions in mouth. He underwent MRI head on 12.01.2023 under sedation. Next day, he was noticed to have sudden onset breathing difficulty with excessive pooling of secretions in mouth. For this illness, he was admitted in GMC, Amritsar for 2 weeks before getting admitted in PGIMER. Parents also reported weight loss of 1.8 kg in last 1 month.

Family history



Examination

HR 142/min, RR 28/min, SaO2 98%, BP 98/46 mmHg, on IPPR
Weight 4 kg (Expected 5.5 kg), OFC 38 cm (Expected 42 cm), Length 60 cm
Chest/ CVS/ Abdomen: Normal

Nervous system

Cranium & spine: Normal; No neck signs
Right convergent squint, weak suck, pooling of secretions in mouth
No ptosis, no facial weakness, No tongue fasciculations
Lying in frog like posture
Bulk: ↓, Tone: ↓ [Appendicular > Axial], Power: 0/5 (proximal), 1/5 (distal),
Deep tendon reflexes: diminished to absent
Plantars: Extensor
Winces to pain

Investigations

	28/01/23	30/01/23	02/02/23	09/02/23	10/02/23	14/2/23	16/02/23
Hb	11.5	**9.3**	**8.2**	**6.9**	10.2	10.6	**8.9**
Platelet	4.7L	4.2L	2.8L	4.8L	3.8L	**91K**	**65K**
TLC	8000	**14600**	13940	12700	**14000**	11500	13500
N/L/M/E	44/49/6/0	**78/17/3/0**	61/21/16/0	77/10/10/1	**74/14/8/1**	83/4/11/0	84/3/11/0

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Na/K	142/4.8	146/3.5	146/3.1	139/4.2	138/49
Ur/Cr	33/0.16	18/0.28	18/0.26	22/0.10	44/0.33
TP/Alb	5.3/4.2	6.0/4.1	6/4.3	**5.1/3.1**	**4.2/2.5**
AST/ALT/ALP	**73/53/60**	**92/116/62**	**141/142/65**	**67/198/60**	**377/96/84**
Ca/P	9.3/3.3	8.8/2.2			8.4/4.7
Bil/Conj Bil	0.54/0.15	0.53/0.15	0.6/0.16	0.4/0.1	0.26/0.08
CRP	**2.44**	**6**	**6.2**	**2.5**	**36**
Procal		**85**			**5.5**

Blood C/S (29/01/23, 30/01/23, 02/02/23, 15/02/23)	Sterile except 1 culture: Staph species **Sensitive to** Ciprofloxacin, Clindamycin, Teicoplanin, Vancomycin, Doxycycline; **Resistant to** Erythromycin, Oxacillin
Central line tip C/S	E.coli **Sensitive to** Tigecycline, **IS** Colistin
ET aspirate	Sterile

Investigations for hypotonia			
CK NAC	**333 (mildly ↑)**	**CK MB**	21 (N)
Serum ammonia	62 (N)	**ECG**	Normal
Lactate	1.1-7 (N/↑)	**Karyotype**	Normal
TMS/GCMS	Normal	**Prolactin**	18.3 ng/ml (N)
TP PCR for myotonic dystrophy	Normal CTG repeat	**Biotinidase**	**3.95 nmol/min/ml (N>5)**

Course and management

The child presented with respiratory distress for which he was started on NPCPAP support. On day 2, he had a cardiac arrest, following which he was started on inotropic support for 48 hours and IPPR for 10 days. He had new onset fever spikes on day 2, for which meropenem was empirically given for 7 days. Then as the child became afebrile with decreasing sepsis markers and sterile blood culture, antibiotics were stopped. He was given T piece trial twice but he did not tolerate due to poor respiratory efforts. He was put on BIPAP support. On day 15, tracheostomy was done. On day 17, he had accidental tracheostomy tube breakage for which bedside tracheostomy reinsertion was done. On day 18, he developed new onset fever spikes with raised inflammatory markers, with worsening pulmonary infiltrates. Keeping possibility of health care associated infection (VAP, fungal sepsis), empirical cefoperazone-sulbactum and fluconazole were started. He continued to worsen with increasing fluffy pulmonary infiltrates. Central line tip culture was positive for E. coli (IS to colistin), hence antibiotics were upgraded to colistin, amphotericin B and oseltamivir. He was started on ventilation with high-pressures. He continued to have refractory hypoxemia day prior to demise with poor peripheral pulses, for which inotropic support was hiked as per protocol. He had cardiac arrest on day 20 and could not be revived.

Throughout the hospital stay, he was evaluated for cause of hypotonia, for which possibility of congenital myopathy, mitochondrial myopathy, spinal muscular atrophy and congenital myotonic dystrophy was considered.

Final Diagnosis

Floppy infant? Congenital myopathy? Mitochondrial myopathy

Cause of death

Suspected Ventilator acquired pneumonia/severe ARDS/catecholamine refractory shock

Attachments:

File:	Size:	Content Type:
CPC Clinical Protocol 07.08.2024.pdf	634k	application/pdf